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Teaching NeuroImage: Nevus Flammeus, Ocular Melanosis, and Seizures in Young Adult With Sturge-Weber and Klippel-Trenaunay Overlap Syndrome

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An 18-year male with delayed gross motor and language development and congenital glaucoma presented with recurrent seizures since three months of age. The Seizures were characterized by left focal onset with secondary generalization and complicated by frequent drug non-compliance. Examination revealed a port-wine stain over the face and extremities (Figure 1A), bluish-grey discoloration of the eyes and periocular region (Figure 1, B, E), and limb length discrepancy with right upper-limb hyperplasia (Figure 1C). These exam findings along with frontal atrophy and gyral calcifications seen on CT (Figure 1D), prompted a diagnosis of Sturge-Weber (SW) and Klippel-Trenaunay (KT) overlap syndrome with ocular melanosis. Both SW and KT syndromes are mesodermal phacomatoses which share port-

wine stain as a common clinical feature¹. The presence of glaucoma, epilepsy, and cerebral gyral calcification are unique to SW while the limb hyperplasia is exclusive to KT¹ with ocular melanosis being additional².

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References

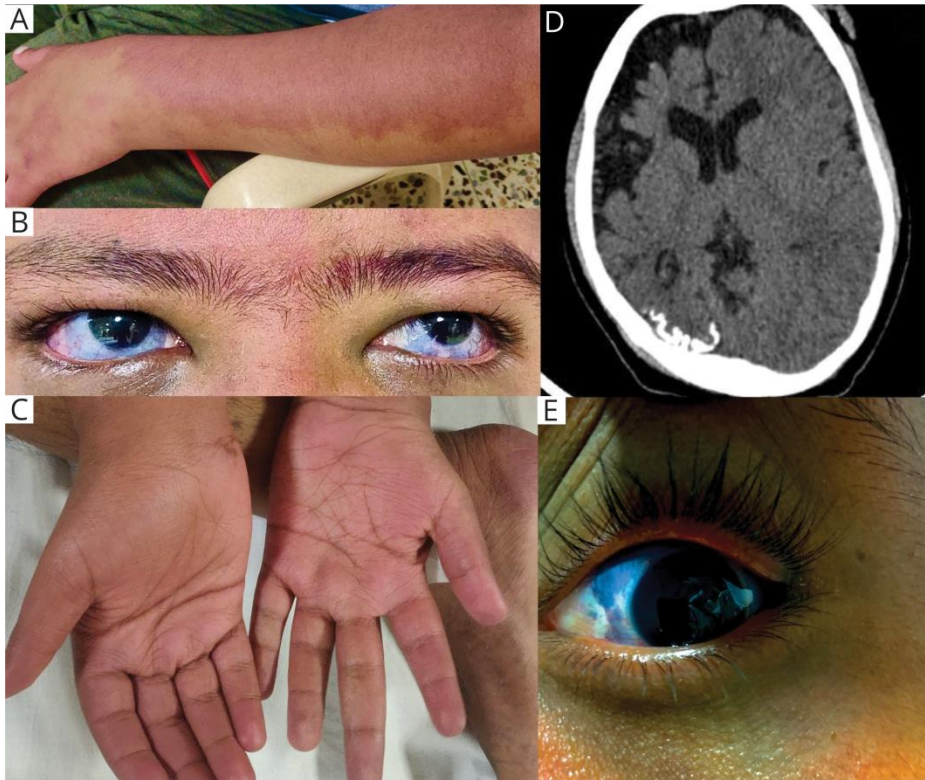
1. Mandal RK, Ghosh SK, Koley S, Roy AC. Sturge-Weber syndrome in association with Klippel-Trenaunay syndrome and phakomatosis pigmentovascularis type IIb. *Indian J Dermatol Venereol Leprol* 2014;80:51-53.
2. Prabhu PB, Soman D, Babitha V. Conjunctival melanosis with retinal vessel tortuosity: An unusual presentation of Sturge – Weber syndrome. *Kerala J Ophthalmol* 2017;29:226-9.

Figure title

Features of Sturge-Weber (SW), and Klippel-Trenaunay (KT) overlap syndrome with ocular melanosis.

Legend

(A) Port-wine stain over the patient's forearm (B) Bluish-grey ocular melanosis (C) Limb length discrepancy with right upper-limb hyperplasia (D) Axial CT image showing right frontoparietal atrophy with gyral calcification in the parietal region. (E) A close-up picture of the patient's left eye



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